

JACKSON (J. B. S.)

(355)

PECULIAR FORM OF SPINA BIFIDA, WITH IMPERFORATION OF THE DUODENUM AND RECTUM.

{Communicated for the Boston Medical and Surgical Journal, Dec. 28, 1858.]

BY J. B. S. JACKSON, M.D.

THE subject of the above malformations was received a few weeks ago from Dr. James W. Robbins, of Uxbridge; and in connection with the case, the following communication, essentially, was made to the Boston Society for Medical Improvement. It was a new-born child, and born apparently at the full period; presenting by the arm, it was turned, and lived about half an hour after its birth. The mother had previously had three well-formed children.

Externally, the penis was quite deficient, and the urethra terminated beneath it and at some distance from its extremity. Over the lower portion of the sacrum was a soft, fleshy excrescence, about half as large as a nutmeg, of a flattened form, and covered by healthy skin. Otherwise well.

The stomach was quite large, and distended. The duodenum terminated in a perfect cul de sac just before the bile duct opened into it, and was distended to the size of a large nutmeg. The intestine, proper, commenced almost at once, and was in no way remarkable except at its lower termination. The rectum opened into the bladder at its fundus, with which it was intimately connected; it then tapered off to a small extremity like an appendix coeci, this terminating portion being about three-fourths of an inch in length, and standing off from the line of the bladder and rectum at a right angle. The bladder was exceedingly small; and the urethra so small, that though the bladder was once, and once only, inflated from it from without, several unsuccessful attempts were made to force air through it by inflating the bladder from the rectum. The right testicle was in the scrotum and the left in the abdomen. Something was also found which it was thought at the time (but erroneously, as the microscope afterward showed) might be an imperfectly developed third testicle, the vas deferens of which joined that of the left testicle about midway. Over the front of the sacrum were two thin, but firm, rounded, well-defined cysts, each one third of an inch or more in diameter; they lay side by side, but the cavities did not communicate, nor were they connected with the neighboring parts except by cellular tissue; these contained apparently a thin serous fluid. The left jugular and subclavian veins formed a trunk that passed down behind and in close connection with the left auricle, and opened into that upon the right side. Otherwise, nothing unusual was observed in the internal organs. The bladder was small, but the kidneys were about as large as usual. Cavity of the pelvis small.

The cutaneous excrescence above referred to, though apparently a small affair, was quite interesting. On removing the surrounding integuments from the parts beneath, a short, rounded, firm,

fibrous-looking cord was seen to run from the excrescence to the termination of the spinal canal. This canal being laid open from behind, it was found that the spinal marrow was continued in substance downward, and nearly or quite through the sacrum, as it is continued down to the sac in a common case of spina bifida when the malformation is below where the cauda equina is usually formed. A small probe passed downward, within the theca, could not be made to enter the cord that seemed to connect the parts within the spinal canal with the cutaneous excrescence. As usually happens in these cases, the posterior laminæ of the last sacral vertebrae were irregularly developed.

In connection with this affection of the spine, I wish to refer to the report of two similar cases in the London *Medical Times and Gazette* (April 17, 1857), in each of which the external tumor, which was of some size, was successfully removed. The writer remarks upon them as "two examples of a condition of things of which, as far as we are aware, no other instances are on record." In the same Journal for April 25th, however, another case is given.

I can hardly believe in the great rarity of this malformation, notwithstanding the remark above quoted, and the fact that it certainly has not been noticed in general treatises, so far as I am aware. During a visit to Europe in 1851, I saw several specimens of this affection, as I regarded them, and of which I made memoranda. One was in the museum at St. George's Hospital, London; finely dissected, and showing the spinal marrow running down in substance into the sac; and in the same museum was a second specimen, unless there was some error in my notes. In the museum of the College of Surgeons, London, is one in an adult subject. In the Hospital Museum at Vienna is a tumor over the back of the sacrum of a new-born child, and which I supposed to be a case of the same kind, though this view of it was not alluded to in the catalogue. At Prague a specimen was seen, and I had some conversation upon the subject with Prof. Engel, to whom the idea of this form of spina bifida was new. At Bologna is a dried foetal skeleton; a cyst, nearly as large as the two fists, arises from the back of the pelvis, and is marked as a subcutaneous encysted tumor. Lastly, in the Hospital Museum at Venice is a solid looking tumor, as large as the head of an adult, and projecting from the back of the pelvis of a nine months foetus.

In addition to the above, I have seen the following cases here; and several of them have been seen by different members of the Society:—

1st. A healthy little girl, partially paralyzed; the tumor looked like a female breast, and a cast was taken of it for the Society's Cabinet (850), in 1845. She is now 18 years of age, and has been able to exercise pretty freely about the city; but for the last few months has been suffering from a pulmonary affection, that her physician, Dr. C. E. Ware, thinks may perhaps terminate in consumption.

2d. An infant, 14 months old, under the care of the late Dr. H. G. Wiley. The tumor was the perfect counterpart of the first case, but there was more paralysis. It died of diarrhoea, in 1843. I examined it after death, and the parts are preserved in the Society's Cabinet (801 and 1223). The spinal membranes expanded into a sac of considerable size, and the spinal marrow was continued down in substance to it. Sacral vertebræ malformed.

3d. In 1846, I received a seven months' fœtus, from Dr. D. H. Storer, and dissected it for the Society's Cabinet (802). This case must have resembled the one seen at Bologna. Instead of a solid tumor, a cyst was found, lined by a polished and vascular membrane, and containing, though by no means distended, ten ounces of serum. The spinal marrow terminated just within an opening upon the inner surface of the cyst. Various other malformations also existed.

4th. A girl, æt. 17, entered the Massachusetts General Hospital, April 3d, 1849, under the care of Dr. J. M. Warren. "The original tumor, directly over the spine (last lumbar vertebra and sacrum), is about as large as a medium-sized apple, is soft and fluctuating, and protected by a thick and tough skin. The second extends from the right side, is about as large as the other, and perfectly solid." I made no record of this case; and, unfortunately, it is not stated in the Hospital Records, from which the above quotation is made, whether there was paralysis; but this last, to some extent, at least, may be inferred from the fact that one of her feet began to turn in when she first began to walk, the other subsequently, and that the deformity was very great when she was seen at the Hospital. The left foot was much diseased, and amputation was performed by Dr. W.

5th. In 1852 I examined, for Dr. J. Homans, a case of extroversion of the bladder with other malformations, in a new-born infant. The tumor of the sacrum was divided into two portions, one of which was half as large as a small orange, and the other equal to a large nutmeg. These communicated freely, contained some ounces of clear serum, and were lined by a serous membrane, between which and the skin that covered them, was a loose cellular tissue, but no fat. Through an opening upon the inner surface of the cyst, a probe was passed upward into the spinal canal; and, the wings of the sacral vertebræ having been cut away, the spinal marrow was found to pass in substance down to the cyst. This case was published in the *American Journal of the Medical Sciences*, January, 1853.

6th. A healthy-looking little girl, æt. $2\frac{1}{2}$ years, under the care of Dr. E. D. G. Palmer. The tumor was nearly or quite half as large as an adult fist, and mostly covered by healthy skin; but from it there arose a mass about the size of a nutmeg, and pretty well defined, of a bright red color, excoriated upon the surface, and having a soft and fluctuating look, as if from a protrusion of

the spinal membranes, though it was quite fleshy to the feel. At another part of the tumor a sort of nævus was seen. The paralysis was very marked, though the child could walk about the room, with the aid of the chairs. The case was published in the Boston Medical and Surgical Journal, May 17th, 1855, and a cast of the tumor, taken for Dr. P. when the child was seven months old, was presented by him to the Society (Cabinet, No. 1284).

7th. A healthy-looking little child, with a solid tumor over the sacrum, and slight paralysis; this case I saw two or three years ago, but made no record of it.

8th. A woman, æt. 23, from St. Albans, Vt., and a patient of Dr. J. L. Chandler. The case was published in this JOURNAL, Feb. 22d, 1855, and with it was given a summary of the four first cases above reported. A cast of the tumor, which was very large and ill defined, is in the Cabinet of the Medical College (1681). The paralysis was considerable, and the feet were much deformed, as in the fourth case; but the general health was good. Recently, I have received a letter from this patient, in which she says that in October, 1857, she fell backward from a horse, struck upon the tumor, and suffered severely in consequence. She is now, however, much better, and the tumor much reduced in size, though still considerably larger than before the accident.

9th. Within a few weeks I have seen, with Dr. Geo. Bartlett, a little girl two years and five months old, in whom the tumor is about one half as large as the patient's head. For the most part the skin is sound, and the mass solid to the feel. To a considerable extent, however, the surface is red and uneven, and the integument inflamed, and so it has been more or less from birth. There is no excoriation of the surface, but a few dry scabs show where there has recently been a discharge, which not infrequently takes place. This discharge is generally purulent, and has sometimes amounted to one or two drachms, but it has never been serious, as if from the spinal canal. Some parts of this red surface are soft, and it has once been punctured, but nothing of any consequence was discharged. In this case there has never been any paralysis.

Dr. Robbins's case, the report of which has led to the above details, makes the tenth and last that I have seen; a sufficient number, certainly, to justify the remark above made, that here at least this variety of malformation is not very rare.

I have always regarded this affection as a form of spina bifida; though I would not undertake to explain why it is that the integument is generally so sound, and the fat so developed beneath it, when the malformation affects the sacral region. I have only once (Dr. H. J. Bigelow's case) seen the healthy skin continued over the tumor when the spina bifida was in the lumbar or dorsal region.

The general characters of this class of cases, as above observed,

may perhaps require some notice. The tumor over the sacrum is congenital, and grows with the subject. The skin and integument are generally healthy, and the whole mass is about as fleshy to the feel and as defined as the female breast. The size varies very much, from a mere excrescence (Case 10) to a very large size. When the skin and integument is imperfectly developed, it is so to a small extent only, in comparison with the whole surface of the tumor. The dilatation of the spinal membranes may be very great, as in the third, the fifth, and in one of the foreign cases; but it is probably and generally very inconsiderable, judging from the feel of the tumor. The spinal marrow is continued down to the sac, so far as dissections have been made; and the sacral vertebrae are probably more or less imperfectly and irregularly developed. The subject, if otherwise well formed, may live to the adult age, but with more or less paralysis; in the ninth case, however, which might be regarded as a formidable one, from the size of the tumor and from the inflammation to a considerable extent of the integuments, this last symptom was wanting.

A very important question arises as to the treatment of these cases. In one of the above, an eminent surgeon proposed to remove the tumor, having probably no idea of its nature and anatomical relations. The patient was thought to have had a narrow escape; and yet when we consider the result of the operation in the two cases reported in the London journal, as above quoted, the fact that the tumor generally grows with the patient, so that in adult life it might get to be very large; and, further, the comparatively small size of the expanded spinal membranes in the large majority of cases, judging from the feeling of the tumor, it would seem very desirable to remove the mass, and possibly the greater part of it might be cut away without opening the spinal cavity. The solid tumor about this cavity probably gives it support, and tends to prevent its enlargement; but support might be given by some artificial means. An opening into the cavity might bring on inflammation that would extend along the spinal and even to the cerebral membranes. In one of the London cases, however, above quoted, and which occurred at the Hospital for Children, the tumor was excised, without the operator seeming to have thought of the spine, and yet the patient did well.

An apology may be necessary for re-publishing some of the above cases which are already in print; but, as so many have now accumulated, I have thought it best to present the whole in a body, and at the same time refer to European specimens, which may be examined by the correspondent of the London *Medical Times and Gazette*, or any others who may feel an interest in the subject.

November 24th, 1858.



